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Endotracheal Involvement as an Unusual Extranodal Site of Recurrence from Mantle Cell Lymphoma

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Mantle cell lymphomas are uncommon subtypes of non-Hodgkin's lymphoma with poor overall survival ranging from 3-5 years. Patients are often middle aged men who would present with generalized lymphadenopathy and typical constitutional symptoms. Extranodal disease is typically found in the gastrointestinal tract, liver, spleen, and central nervous system. Lung involvement is typically restricted to parenchymal lesions. Endotracheal involvement is quite uncommon with less than 5% of cases. We present an unusual case of treated mantle cell lymphoma recurring 7 years later with only endotracheal involvement demonstrated by fluorodeoxyglucose-positron emission tomography.

Introduction

Mantle cell lymphomas are uncommon subtypes of non-Hodgkin's lymphoma with poor overall survival ranging from 3-5 years. Patients are often middle aged men who would present with generalized lymphadenop-

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Abbreviations: CT, computed tomography; FDG, fluorodeoxyglucose; PET, positron emission tomography

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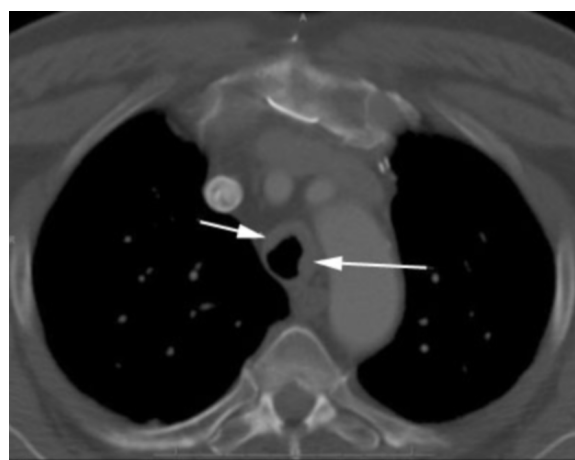


Figure 1. 55-year-old man with recurrent mantle cell lymphoma. CT shows symmetric circumferential thickening of the distal trachea (arrows).

athy and typical constitutional symptoms. Extranodal disease is typically found in the gastrointestinal tract, liver, spleen, and central nervous system. Lung involvement is typically restricted to parenchymal lesions. Endotracheal involvement is quite uncommon with less than 5% of cases. We present an unusual case of treated mantle cell lymphoma recurring 7 years later with only endotracheal involvement demonstrated by fluorodeoxyglucose-positron emission tomography.

Case Report

A 55-year-old man presented to the emergency room complaining of left lower quadrant abdominal pain, abdominal distention and nausea. Review of systems revealed a 20-pound weight loss over four months and frequent night sweats. No prior history of malignancy. His mother died of Hodgkin's lymphoma. Patient has a history of coronary artery disease status post angioplasty, hypercholesterolemia, and hypertension. Physical exam revealed a markedly enlarged tender spleen and inguinal lymphadenopathy. Lab work revealed anemia, leukocytosis and borderline thrombocytopenia. CT scan was performed which demonstrated splenomegaly. Lymph

node and bone marrow biopsy was performed which revealed non-Hodgkin's lymphoma, mantle-cell type.

Patient was then started on chemotherapy using the CHOP protocol for six cycles. Rituxan was added 6 months later after a partial response. Patient later received an autologous stem cell transplant in March 2001. Patient also underwent splenectomy after transplant. Patient remained in remission until follow up CAT scan performed in January 2008 which demonstrated circumferential thickening of the distal trachea (Figure 1). 18F-FDG PET scan was performed a month later demonstrating avidity only within the distal trachea (Figures 2 and 3). Bronchoscopy and biopsy was performed which was positive for mantle cell lymphoma. Patient refused surgery and was then referred to radiation oncology for up to 4000 cGy delivered to the distal trachea.

Discussion

In 2004, more than 56,000 cases of non-Hodgkin's lymphoma occurred in the United States with an incidence of about 22 per 100,000 and a death rate of about 8.8 per 100,000. Mantle cell lymphomas represent

between 4-8 percent of non-Hodgkin's lymphomas. Older individuals are more affected with the median age of diagnosis of 63 years. The male to female ratio is about 4 to 1 [1, 2].

Diagnosis primarily rests upon biopsy of an affected lymph node or extranodal site of disease. Specific classification relies upon the specific macro and microscopic appearance in addition to its immunological phenotype and genetic abnormalities. CD markers include: CD20+, CD3-, CD10-, CD5+, CD24-, and PRADI+. The most frequent cytogenetic abnormality would include t(11;14) (q13;q32). Mantle cell lymphoma

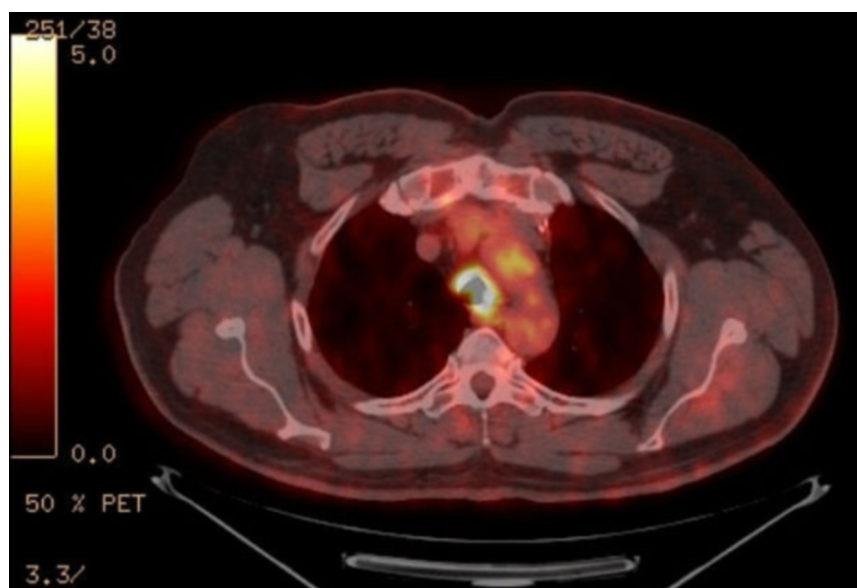


Figure 2. Axial PET/CT shows intense activity in the suspected area around the distal trachea.

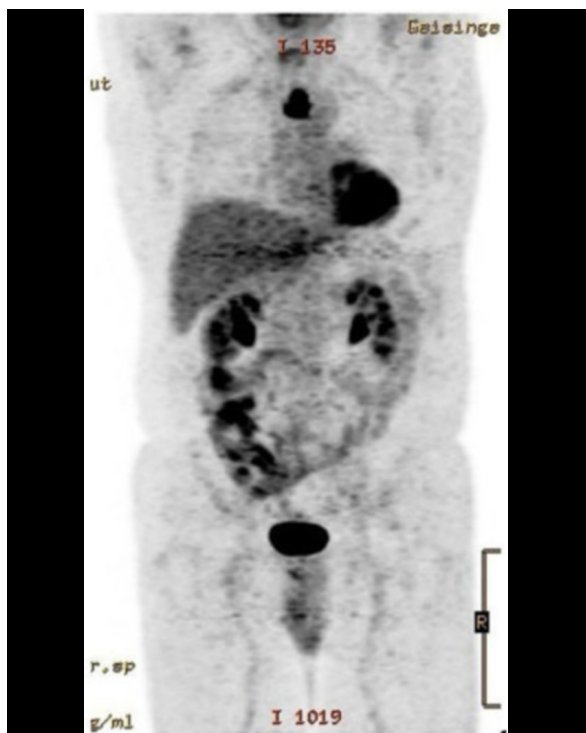


Figure 3. Full body coronal reformatted PET scan shows abnormal intense tracheal uptake and other areas of expected uptake.

phomas are associated with the bcl-1 oncogene [1].

Mantle cell lymphomas are frequently diagnosed, over 70% of the time, with stage IV disease involving more than one extranodal site in 50% of cases. Clinically, again generally being older men, will present with generalized lymphadenopathy, sometimes massive splenomegaly and diffuse GI polypoid lesions in addition to the typical “B” symptoms [1]. Mantle cell lymphomas are considered to be an indolent lymphoma variant with patients surviving many years without treatment. Aggressive disease will usually get a standard CHOP-R regimen. Patients will usually experience a very high overall response rate to induction treatment with a short time to progression. Overall survival is about 3–4 years [1].

Extranodal sites of non-Hodgkin’s lymphomas commonly involve the gastrointestinal tract, liver, spleen, central nervous system, bone marrow, pulmonary

parenchyma and pleura. Other sites account for less than 5–10% such as kidneys, adrenals, testes, bone, head and neck. Rarely will muscle, breast, pancreas, and the trachea be involved [3].

CT has been the gold standard for staging, however is limited to only structural detail. Extranodal spread can be missed if not pathologic by appearance. For initial staging, studies have shown that upstaging occurs in about 20% of patients when 18F-FDG PET is added. Aggressive disease can also be determined by SUV uptake greater than 10. PET will also add information about treatment response and prognosis. Combined PET/CT can increase sensitivity and decrease false positives partially when brown adipose tissue mimics disease [4].

Our patient’s CT at interval follow-up, prompted addition of PET to confirm disease versus other benign entities (e.g. endotracheal mucus). CT alone could have delayed treatment. For the time interval and site of recurrence, this case is quite unusual.

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